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IS EPILEPSY A FUNCTIONAL DISEASE?[1]

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In our conception of diseases tradition plays an important part and often handicaps investigation. In medical text-books from the earliest times epilepsy has been classed as a functional disease, and this view does not seem to have been seriously questioned. Yet a careful review of many facts offered by recent advances in our knowledge of nervous affections seems to me to demonstrate the fallacy of the prevailing opinion and to prove conclusively that epilepsy is usually, if not always, an organic disease.

These facts I wish to present to your consideration in the hope that when they are criticized and discussed we may come to an agreement and thus change permanently the conception which we have hitherto entertained in regard to the nature of epilepsy.

My argument will not be a theoretical one only. It is based upon a careful study of 2,000 cases of epilepsy, which have been seen personally, and of which I have satisfactory records. I shall not, however, weary you with statistics drawn from these cases. They are presented in an appendix to this paper in the form of tables. I shall, however, draw upon these tables for a number of statements which they prove.

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[1] Read before the Philadelphia Neurological Society, Nov. 24, 1903.

A. In the first place, I think we all agree that it is possible to draw a fairly sharp line clinically between Jacksonian epilepsy and ordinary so-called idiopathic epilepsy.

The recognition of Jacksonian epilepsy is now universal, and we can distinguish several distinct types of this affection:

(1) There is the motor type, in which a local spasm of one or more parts upon one side of the body only—followed by temporary weakness—is the characteristic;

(2) There is the sensory type, in which a sudden hallucinatory perception occurs in any one of the many senses, to be followed by a temporary suspension of the power of perception in that sense;

(3) There is the aphasic type, in which a sudden interference with the function of speech takes place, either in its receptive or on its emissive side—either in the power of understanding or in the power to speak;

(4) There is the psychical type, in which dreamy states of the mind or imperative ideas dominate consciousness, arresting the normal flow of thought, and often leading to automatic acts whose object is not clear and of which no conscious memory remains.

In all these types we consider the Jacksonian attack a sure indication of local irritation of the brain cortex; we admit that it is a symptom of organic disease; we are guided by the variety of the attack to locate the disease and to proceed on the basis of the localization to operative interference in many cases. It needs no argument to-day to support the position that organic disease is the cause of Jacksonian epilepsy.

The form of organic disease may be one of many kinds. Thus tumors, abscesses or clots on or near the cortex are frequently productive of Jacksonian epilepsy. A focus of softening or of sclerosis due to embolism or thrombosis in a blood vessel, or due to a syphilitic deposit in the vessel wall or in the membranes or in the brain substance may be a cause. A placque of gliomatous infiltration, such as we find in multiple sclerosis, may be a cause. But, whatever the organic lesion, the effect will be the same, provided it produces an irritation of the brain cortex.

It is not at all surprising that in any case of Jacksonian epilepsy the character of the attack is uniform. The fit always begins in the same way in the individual affected. This is because the disease present has a definite location and produces a uniform kind of irritation in a single focus.

The attack is always recognized by the patient as one of a similar series, and, as consciousness may not be lost, the patient's description of the attack may be accepted.

It has been stated that a sharp line is drawn clinically between Jacksonian epilepsy and idiopathic epilepsy.

Let us see whether such a sharp distinction is justified:

In many cases of ordinary epilepsy (38 per cent. of my cases) the attack is preceded by a conscious sensation which we call an aura. That aura is in many cases identical in character with the sensation constituting or starting the Jacksonian attack. It is always uniform in the same patient, as are Jacksonian attacks.

The only difference between a Jacksonian attack of the motor type and an idiopathic attack is the extent of the spasm. In one it is limited, in the other general. In one it begins in one part and extends; in the other it begins in many parts, at once. In a Jacksonian attack the spasm may extend so as to finally involve all the limbs, and the Jacksonian attack may run into a general convulsion. In both a state of motor weakness follows the attack. Thus there is no essential difference between the two.

The same may be said of the sensory type. In the Jacksonian attack there is a sensation which often goes on to a spasm, and this in turn may go on to a convulsion. In ordinary epilepsy an aura which is identical in many ways with a Jacksonian sensory attack occurs and goes on to a convulsion. Thus in my table of varieties of aura I find numbness in one arm or both in 76 cases, numbness in one leg or both in 27 cases, numbness in one or both sides of the face in 22 cases, numbness in the trunk in 4 cases.

Had these patients had a local spasm and not a general convulsion after the aura we should at once have suspected a lesion in the sensori-motor area.

I find visual hallucinations in 68 cases, auditory hallucinations in 30 cases, hallucinations of smell or taste in 14 cases.

Had these patients not had a general convulsion following we should have suspected a lesion in the occipital, or temporal or uncinate convolutions respectively.

I find in many cases that the fit begins with a cry—a discharge from the motor speech centre. I find in 57 cases an aura of a mental kind, a fear, a sense of bewilderment, a forced impulse to say and do something, a condition quite analogous to the psychical state seen in the Jacksonian attack, only differing from it in its sequel of a fit. I find in 158 cases a sense of vertigo, a loss of equilibrium, a sensation so frequently met with and so characteristic of lesion of the cerebellum that one might almost classify these cases as cerebellar epilepsy. It is possible that the sudden fall occurring in many cases is due to a sudden suspension of the cerebellar function.

Is there any sharp line to be drawn between Jacksonian epilepsy and ordinary epilepsy? Is it not merely a difference of degree in attacks essentially similar? Is it not probable that when the difference is so slight the underlying cause may be the same? Is it not possible that whenever we have a general convulsion preceded by a definite form of aura, we can conclude that we are dealing with a case in which a focus of irritation, organic in nature, is present in a definite locality in the brain? I see no other explanation for the attacks of epilepsy preceded or attended by a uniform aura. And this class amounts to 23 per cent. of my 2,000 cases.

It may be said that a sharp line has been drawn between Jacksonian epilepsy and ordinary epilepsy, on the ground that in the former there is no loss of consciousness, while in the latter there is such a loss. I think the difference here lies again in the degree of severity and rapidity of extent of the cerebral irritation and consequent shock. If a Jacksonian attack becomes very severe and widespread, consciousness is often lost. Any sudden shock to the brain obscures consciousness for a time. The time may be but a fraction of a second, as after a sudden blow on the head or a petit mal attack. It may be longer, as in a grand mal attack. It may be incomplete, as in those attacks in which a patient does not fall, but balances himself and makes inarticulate noises and slight grimaces, but is not convulsed and is merely bewildered. In this respect the difference between an attack, without or with a loss of consciousness, does not appear to argue any underlying difference in conditions present.

Therefore, the admitted fact that Jacksonian epilepsy is due to organic disease and the close resemblance in many cases (about 23 per cent.) between the attacks of Jacksonian and ordinary epilepsy seem to me to offer an argument that, in these cases at least, the ordinary type is due to actual lesions.

B. The second argument for the organic nature of epilepsy is drawn from a study of cases of maldevelopment of the brain. These cases are not included in my table of 2,000 epileptics, because I do not classify them together. But any one who has studied the subject of maldevelopment of the brain in the clinical types presented so commonly, viz., (1) imbecility and idiocy; (2) hemiplegia of infancy; (3) diplegia of infancy, or Little's disease; (4) the sensory types of infantile cerebral paralysis, cannot but admit that in these conditions epilepsy is a most common complication.

Of 400 cases of maldevelopment of the brain elsewhere analyzed[2] 156 patients were subjects of epilepsy, i. e., 39 per cent. Many of these patients presented such manifest symptoms of mental and physical defects that the epilepsy was admitted to be a secondary and minor matter. But it is not at all uncommon to have a child brought who is supposed to be suffering from epilepsy only; and to find, on obtaining a history or on making a physical examination, that, prior to the development of epilepsy there has been an apoplectic attack which has left a trace of hemiplegia or hemianesthesia or hemianopsia or mental weakness. In many cases the history shows that difficulty at the time of birth had resulted in a state of asphyxia or a slight birth palsy of cerebral origin, which had attracted only a little notice, as it had subsided, but later epilepsy has developed without known cause. (38 cases, 2 per cent.)

In two patients, brought to me for epilepsy only, I have found bilateral homonymous hemianopsia which had been congenital, and neither of these children was aware that vision was defective. In both a history was obtained of hard labor with forceps delivery and also difficulty in maintaining the life of the infant for some days. In both the epilepsy had developed during the second year, but in neither had any defect of the brain been suspected until my examination showed the hemianopsia. In both a flattened skull in the occipital region indicated a defective development of the occipital lobe. Here was a clear proof that a congenital lesion had been the cause. Among my 2,000 cases of epilepsy, 192 showed positive signs of maldevelopment of the brain.

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[2] Organic Nervous Diseases," page 530.

In the cases of maldevelopment of the brain suffering from epilepsy, the epileptic attack may be Jacksonian in type, or it may be an ordinary fit. It may be preceded by an aura of sensory or motor type, or it may not, though in this class of epileptics the presence of an aura has been noticed in a much larger percentage than in idiopathic epilepsy.

If we study the age at which idiopathic epilepsy develops, we must confess that it is a disease of childhood and youth. Of my 2,000 cases, 622 developed before the age of 10, and 755 between the ages of 10 and 20; that is, 68 per cent. developed before 20 years of age. The disease is one, therefore, which appears during the period of brain development.

When epileptics are carefully studied, it is usually possible to find evidences of some mental or physical sign of degeneracy or of defective evolution, and this is particularly true of those who are affected in infancy or in youth.[3] The difficulty experienced by parents and teachers in educating epileptic children is not merely to be traced to the irregular attendance upon school, due to the occurrence of fits, nor to the depressing effect of bromides on mental activity and memory. It is due much more commonly to an inherent deficiency in the mental power, especially in those higher powers of the mind—attention, concentration, and self-control.

Do not these facts then suggest strongly that, as epilepsy is a symptom of organic disease in the cases of maldevelopment of the brain, it may also be a symptom of organic disease in those cases in which its appearance in early life and its association with imperfect capacity suggest an imperfect brain growth. Are there not many cases, supposedly idiopathic and without apparent cause, which are actually due to some slight and easily overlooked defect in the brain? And if, in such cases, the epilepsy is a mere symptom of brain defect, these cases are rather to be classed with the cases of defective mental development of organic origin than to be classed with the functional neuroses.

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[3] A somewhat arbitrary classification of the mental state was attempted in the study of my 2,000 cases. It is recorded as idiotic in 11 cases; imbecile in 16 cases; poor in 580 cases; fair in 269 cases; good in 854 cases. Thus in about 44 per cent of the patients there was some evidence of defective mental power.

And this leads me to remark that it is a fallacy to state that an epileptic presents no symptoms between the attacks. This statement is often made and conveys the idea of a disease characterized by occasional attacks, but without permanent continuous symptoms—like a migraine.

In my experience it is the exception rather than the rule to find an epileptic in a perefctly normal mental and physical condition without signs of degeneracy or symptoms of some imperfect function. Many patients show such signs and symptoms. They may not be noticed by the parents. They are often recognized by the physician. They are sometimes wrongly interpreted as the cause of epilepsy. Much futile treatment of epilepsy has been based upon the discovery of defects of growth or structure or function, and upon attempts to correct these, while the eyes have been closed to the fact that such defects and the epilepsy have no causal relation, but are both effects of an underlying condition. Here come in the cases of so-called reflex epilepsy, where an attempt is made to cure an underlying organic disease of the brain by cutting eye muscles, removing adenoids or tonsils, washing out the stomach or colon, circumcising and removing ovaries or correcting uterine displacements. I do not deny that it is our duty to remove all causes of irritation from a nervous system peculiarly sensitive to peripheral impressions and peculiarly liable to react to such impressions by a nervous explosion. But I do deny that such measures can cure epilepsy, for I do not believe that peripheral irritation ever actually caused the disease. I am constantly seeing cases where, in spite of such treatment, the disease persists.

Therefore I would maintain that many cases of idiopathic epilepsy are actually cases of maldevelopment of the brain, with gross or microscopic defects; that the associated mental and physical signs of degeneracy merely indicate this maldevelopment, and that, like them, the epilepsy is a manifestation of an organic disease, or defect.

C. Another argument for the organic nature of epilepsy can be urged from a study of the alleged causes of the affection. (See Table V.) I use the term supposed causes, for it is not always certain that an event or a condition preceding the development of the disease has a causal relation to it.

The first of these causes is heredity. In my 2,000 patients there was supposed to be a bad inheritance in 700 cases. By bad inheritance I mean the existence of epilepsy in some near relation (136 cases), of alcoholism in a parent (120 cases), of serious nervous disease (118 cases), of rheumatism (108 cases), of tuberculosis (76 cases), or of some combination of these (143 cases). (See Table IV.) This does not imply that the disease itself is inherited or transmitted. Were it so the proportion of cases of direct transmission would be much larger (17 cases in 2,000), and, as one person in 500 in the community is afflicted by the disease, the increase in its frequency would be far greater. It does imply that the patient inherits a weak and probably a defective nervous system—has, in fact, an organically imperfect brain.

The second of these causes in frequency is trauma of the head. That trauma of the head is more commonly followed by epilepsy than trauma of other parts of the body is proved by the statistics of the Franco-Prussian war. The records show that, among 8,985 persons wounded on the head, 46 developed epilepsy, while among 77,461 persons wounded elsewhere only 17 became epileptic. I have a history of trauma in 229 cases, i.e., 11 per cent. When a fracture of the skull causes a laceration of the brain, we are never surprised at the subsequent development of epilepsy, and we never question the organic basis of the disease. In many cases there is no apparent fracture of the skull, but there is every reason to suppose that a hemorrhage within the brain has occurred—very possibly small in extent and capillary in origin—producing, however, a destruction of tissue; which is followed by the formation of a scar in no way different from that produced by an accidental or a surgical wound.

The traumatic cases, therefore, are in all probability dependent upon an organic lesion in the brain.

In the list of supposed causes tabulated are many of the infectious diseases—scarlet fever, measles, whooping-cough, diphtheria, typhoid fever. Marie has shown that multiple sclerosis of the nervous system—a disease in which epileptiform attacks may occur as a symptom—is probably a sequel of these diseases and due to the infectious agent, microorganism or toxin, which causes them. Is it not possible that in the epileptic cases developing after an infectious disease some similar sclerotic plaque may have been produced in the brain, and that in these cases the disease has an organic basis? Among my 2,000 cases, 50 developed as a sequel of such diseases.

The cases following sunstroke are undoubtedly organic in origin and due to capillary hemorrhages.

The epilepsies of advanced life, which are happily few in number (but 26 cases developed after the age of 50) can almost invariably be traced to endarteritis and atheroma. This is proven by the fact that, as a rule, these patients have attacks of apoplexy, indicative of cerebral hemorrhage or thrombosis, either just before or soon after their first epileptic fit. Hence we may believe that senile epilepsy is surely of organic origin.

Other supposed causes, such as childbirth, dentition, syphilis, menopause, overwork, etc., seem to me too vague and uncertain to be accepted as true etiological factors, and do not throw any light on the pathology. Fright is undoubtedly a cause, as 119 patients had the first fit soon after a fright, too large a number to be explained as mere coincidence. But this, again, has no known bearing upon pathology.

Therefore the study of the supposed causes indicates that in the cases in which they can be accepted as probable causes, they are so because they have produced organic disease of the brain.

From a consideration of these various lines of argument it seems to me probable that epilepsy is to be regarded as an organic disease.

D. This is not a new idea. The literature of epilepsy from the time of Schroeder van der Kolk to the latest treatise by Binswanger, is full of observations in its confirmation. But the stumbling block in the way of their acceptance has been that each observer in turn found some new lesion or located the disease in a new region of the brain. And as the tendency of the scientific mind is to search for a single cause for an effect, the multiplicity of causes advanced by pathologists was thought to detract from the force of their conclusions. The sclerosis of Ammon's horn noticed by Meynert and also found by many others has been forgotten or passed over. The gliosis disseminata found by Féré, Chaslin, Alzheimer and Eurich has received little attention. And the recent studies by L. Pierce Clark and Prout and others of the finer degenerative changes in the cells of the cortex found by them uniformly in the brains of epileptics have excited little interest. As a matter of fact, all these observations are worthy of most careful attention. They all demonstrate the existence of gross or microscopic changes in the epileptic brain.

Conclusion.—The brain is a storehouse of energy, ready to give it forth in orderly manifestation as in voluntary action and speech; or in disorderly discharge, as in an epileptic attack. The attack is the symptom, it is not the disease. The disease is a disorder of control over inherent energy. Its existence is proof of a weak and defective organization of the brain, and any lesion, no matter of what kind, and no matter in what place, is capable of interfering with these mechanisms of control and of giving rise to the symptom. When the symptom is present there is evidence of a defect of control. And it seems to me far more reasonable, in the light of all the facts here presented, to admit that epilepsy is an organic disease of the brain characterized by a lack of control over the mechanisms of motor energy.

TABLE I.

Age at onset.

Under	Cases	Under	Cases	Under	Cases	Under	Cases
1 yr.	67	11 yrs.	74	21 yrs.	66	31 yrs.	42
2 yrs.	93	12 “	62	22 “	61	32 “	14
3 “	80	13 “	77	23 “	53	33 “	18
4 “	55	14 “	74	24 “	37	34 “	18
5 “	58	15 “	114	25 “	28	35 “	13
6 “	49	16 “	85	26 “	26	36 “	14
7 “	53	17 “	92	27 “	28	37 “	11
8 “	60	18 “	71	28 “	29	38 “	11
9 “	47	19 “	64	29 “	17	39 “	13
10 “	56	20 “	42	30 “	23	40 “	6
	618		755		368		160

Under	Cases	Under	Cases	Under	Cases
41 yrs.	12	51 yrs.	3	63 yrs.	2
42 "	6	52 "	3	66 "	1
43 "	8	53 "	2		
44 "	4	54 "	2	70 "	1
45 "	7	55 "	1	74 yrs.	1
46 "	6	56 "	0	76 "	1
47 "	9	57 "	1		
48 "	4	58 "	2		
49 "	5	59 "	2		
50 "	2	60 "	2		
	63		18		6

Males 1163. Females 837.

TABLE II.

Character of Attacks.

a

	Cases
Grand mal	1125
Grand and petit mal	580
Petit mal	170
Jacksonian	37
Psychic	16
Grand mal and Jacksonian	10
Grand and petit mal and Jacksonian	8
Grand mal and psychic	3
Grand mal petit mal and psychic	6
Petit mal and psychic	2

b

	Cases
Diurnal and nocturnal	882
Diurnal only	658
Nocturnal only	380

TABLE III.

The Varieties of Aura.

An aura was present in 38 per cent of the cases, and in almost every patient never varied in its character in different attacks. The following varieties of aura were noted:—

	Cases
Epigastric sensation	198
Cephalic sensation of vertigo	158
Numbness in the arms	76
Visual hallucinations	68
Mental states with fear	51
Cardiac sensations	34
Auditory hallucinations	30
Abdominal sensations	25
Numbness in the legs	27
Numbness in the face	22
Respiratory sensations	16
Hallucinations of smell	8
Hallucinations of taste	5
Sensation of numbness in trunk	4
Sensation of thirst	4
Hallucinations of smell and taste	1
Sensation of hunger	1
Sialorrhea	1
Diarrhea	1
Tremor	1

TABLE IV.

Hereditary factors noted in 700 cases.

	Cases
Epilepsy	136
Alcoholism	120
Nervous diseases	118
Rheumatism	108
Tuberculosis	76
Alcoholism and rheumatism	38
Rheumatism and nervous diseases	18
Tuberculosis and nervous diseases	19
Rheumatism and nervous disease	18
Tuberculosis and rheumatism	16
Alcoholism and tuberculosis	16
Alcoholism and epilepsy	7
Alcoholism, nervous disease and tuberculosis	5
Rheumatism and epilepsy	3
Alcoholism, rheumatism and tuberculosis	2

TABLE V.

Alleged Cause of the Epilepsy.

	Cases		Cases
Trauma	229	Sunstroke	12
Fright	119	Physical strain	11
Alcoholism	61	Measles	12
Invalidism	49	Indigestion	10
Mental strain	36	Arterial sclerosis	8
Menstrual disorder	24	Syphilis of the brain	8
Dentition	23	Whooping cough	6
Child-birth	24	Rickets	5
Scarlet fever	20	Excessive heat	5
Infantile palsy	19	Trauma to mother	4
Maldevelopment of brain	19	Diphtheria	4
Masturbation	14	Typhoid fever	4
Menopause	11		